Case Report

Myxopapillary Ependymoma: A Short Compilation with a Case Report

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ABSTRACT

Myxopapillary ependymomas (MPEs) are a variant of the spinal ependymomas, which are common in the conus medullaris and filum terminale in the sacrococcygeal region. Approximately 13 % of the spinal ependymomas are myxopapillary ependymomas. These tumors are usually a gradual development. It is mostly localized in the intradural space and may cause local pressure on the paravertebral soft tissue and neighboring bone. MPEs may be found in extraspinal, extradural, subcutaneous tissues, in the cervical and thoracic vertebras and in the brain. The prognosis of the small lesions are usually good and a complete resection is usually curative. A local extension to the soft tissue and a dissemination to the subarachnoidal space may be encountered as a result of the incomplete resection. Our objective was to remind this rare tumor with the occasion of an adult patient.

INTRODUCTION

Ependymomas are the most common primary intramedullary spinal cord tumors and consist 30-40 % of the cases.(1, 2) Although they may be encountered in every age group, they peak between the ages of 0-4 years and between 55-59 years (3).MPEs, which are a subgroup of the spinal ependymomas, are usually localized in conus medullaris and filum terminale in the sacrococcygeal region.(4). MPEs were first described by Kernohan in 1932.(5) Ependymomas were categorized by World Health Organization (WHO) in 3 grades: Grade 1: myxopapillary ependymoma and subependymoma; Grade 2: ependymoma (conventional) and Grade 3: anaplastic ependymoma. Most of the lesions are in Grade 1 in the clinical practice.(6)

CASE REPORT

The 34-year old male patient applied to our clinic with the complaint of lumbar pain for the last two months. There was no trauma in his medical history. His neurological examination was normal. The lumbosacral region was examined with the magnetic resonance imaging(MRI).(Figure 1) An intradural, extramedullary contrast-enhancing lesion (diameter: 13 mm), which applied pressure on the cauda equina, was observed at the L_1-L_2 level and caudaequina in the central canal. The patient was referred to the neurosurgery clinic and underwent intradural total lesion excision. The examination of the pathological specimen showed a Grad 1 myxopapillary ependymoma with a chi-index of 6-7 %. The immunohistochemical staining displayed S100, Vimentin and GFAP expression in the tumor cells. Postoperative contrast-enhanced spinal MRI and cranial MRI evaluation were normal. The cytological examination of the craniospinal fluid was negative. The patient was followed up for one year at 3-month intervals.

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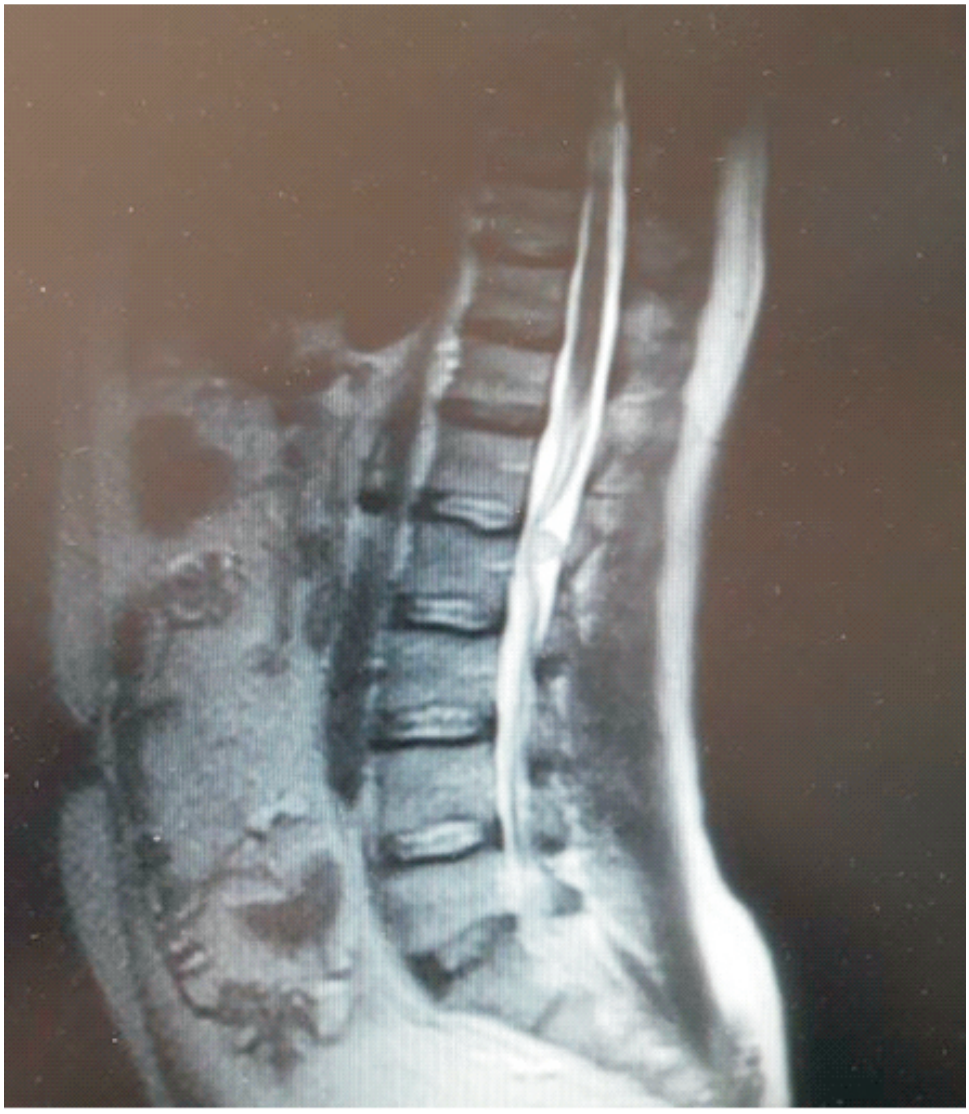

Figure 1. Sagittal magnetic resonance imaging of the lumbar spine showed the primary lesion at L_1-L_2 level (black arrow)

DISCUSSION

The most common symptoms of MPE are a lumbar or sacral pain. Approximately 25 % of the patients have weakness in the lower extremities and sphincter dysfunction. An intradural extramedullary localization is common and rarely localized in the extradural space. Compared to other spinal ependymomas, MPEs emerge in earlier ages and the mean age is 35 years. Expansion to the craniospinal fluid may be observed.14-43 % of the cases have multiple lesions. MPEs are GFAP, S100 and vimentin-positive. Histopathologically, MPEs consist of papillae characterized by the myxoid degeneration and hyalinized blood vessels.(7) Regarding the differential diagnosis of MPE, paraganglioma, chordoma, neurofibroma, extraskeletal chondrosarcoma, metastatic papillary carcinoma and metastatic carcinoma should be considered.

In a study, 282 female and male ependymoma patients, who had applied to 19 different health centers, were investigated. Their mean age was 43 years (18-80 years). In 129 patients, the lesions had a spinal localization (46 %). According to the WHO categorization, 78 % of the patients were in Grade 2, 11 % in Grade 1 and 11 % were in Grade 1. All Grade 1 tumors were myxopapillary ependymoma. The cases with spinal localization had the highest 5-year survival rate (97 %).(8)

5-year survival rate was 98 % in cases, who underwent total excision. As local recurrence may be observed in cases with subtotal excision, a postoperative radiotherapy is necessary.In the presence of an expansion to the subarachnoidal space and involvement of the root of cauda equina, resection is usually incomplete and the rate of the local recurrence is increased. Although the recurrence risk is low in MPE, a long-time follow-up is required and distant metastasis may be observed in rare cases (e.g. lung).

CONCLUSION

MPE is a rare variant of ependymomas. They have a low risk of recurrence and a long follow-up period is necessary. Total excision of the primary tumor is the most important factor in the treatment and the prognosis. Radiotherapy should be added to the treatment in cases with an expansion to the surrounding radices and subtotal resection.

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